Esophageal Atresia and Tracheoesophageal Fistula in the Twin

Anatomic Variants

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Recent experience with a twin who had esophageal atresia and tracheoesophageal fistula revealed complex anatomy not suspected before operation. The experience in patients with esophageal atresia and tracheoesophageal fistula who were twins was reviewed at the Children's Hospital of Los Angeles. Of 245 patients seen in the past 23 years, 16 were twins (only two of whom were siblings). Six of the 16 patients (38%) had other than the most frequent anatomy, *i.e.*, proximal atresia of the esophagus and distal fistula. Two of these patients had extremely complex anatomy. Twenty-five per cent of the patients had a right aortic arch compared with 5% of all patients with tracheoesophageal fistula. The surgeon who encounters a twin with esophageal atresia or tracheoesophageal fistula is appropriately cautioned that significant anatomic complexity may be encountered.

THE INCIDENCE OF twinning in patients with esophageal atresia and tracheoesophageal fistula is essentially the same or just slightly greater than in the general population.[1] Recently, we operated on a twin with tracheoesophageal fistula who had an unusually large fistula associated with a laryngotracheoesophageal cleft. This patient prompted a review of experience with esophageal atresia and tracheoesophageal fistula in twins at the Children's Hospital of Los Angeles.

Methods

All patients admitted to the Children's Hospital of Los Angeles from January 1960 through December 1983 with a diagnosis of tracheoesophageal fistula and/or esophageal atresia were reviewed. In this group there were 16 twins, 14 of whom had a normal sibling/twin, and one set of monozygotic twins, both of whom had esophageal atresia and tracheoesophageal fistula.

The 16 charts were reviewed specifically for the following data: sex, birthweight, type of anomaly, associated congenital anomalies, operative procedures performed, and results, including morbidity and mortality. The side of the aortic arch was recorded for each patient. The surgeon's dictated operative note was used for a description of the anatomy.

Survivor follow-up data were obtained from clinic records.

Results

The classification used to describe the anatomy of the congenital defect is descriptive and does not follow a preset lettered system. This anatomic classification is found in Table 1.

It should be noted that the two twins with laryngotracheoesophageal cleft were the only patients with this rare problem encountered at the Children's Hospital of Los Angeles over the past 25 years. The monozygotic twins had proximal esophageal atresia and distal tracheoesophageal fistula without other significant associated anomalies.

Table 2 is a compilation of the anomalies discovered in this series. Many of the patients were found to have multiple anomalies.

Discussion

The largest published series of infants with esophageal atresia and tracheoesophageal fistula is the report of the Surgical Section of the American Academy of Pediatrics (AAP) (published in 1964) which described 1054 cases.[2] Statistics from other large series on anatomic details have in general correlated closely.[3,4]

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Submitted for publication: September 18, 1986.

Reports of twins with esophageal atresia are relatively rare but can be found sporadically over the last 25 years.[5–8] As far back as 1670, there is a description of tracheoesophageal fistula occurring in conjoined twins. German et al.[9] reported on twins with tracheoesophageal fistula at the Children's Hospital of Los Angeles, focusing primarily on survival as related to weight.

Our series demonstrates almost three times as many instances of uncommon anatomy among twins with esophageal atresia as that found among nontwins. In the AAP survey, 87% had proximal atresia and distal tracheoesophageal fistula, compared with 63% in our series. Also, two of those six patients with uncommon anatomy had a very complex laryngotracheoesophageal cleft. One of these two patients was one of the first successful repairs of a cleft reported in 1973.[10]

Several individuals have commented on the association of right aortic arch and tracheoesophageal fistula. This association can lead to significant technical difficulty if the fistula is approached through the right chest, such that the procedure may need to be abandoned and subsequent repair undertaken *via* the left chest, with attendant increased morbidity, length of hospitalization, and, therefore, cost. The usual incidence of a right aortic arch in the patient with a tracheoesophageal fistula is 5%,[11] but one fourth of our patients manifested this association, thus making it even more crucial to ascertain the side of the arch before thoracotomy. This can be done in several ways. We have conventionally placed an umbilical artery line and then confirmed the side of the arch by plain film, but with increasing experience our cardiologists have successfully obtained the same data by noninvasive cardiac ultrasound.

Because of the timing in embryogenesis at which esophageal atresia with tracheoesophageal fistula occurs, it is the exception rather than the rule that this should occur as an isolated defect. The incidence of associated anomalies ranges from 50–70%.[2,4] Our series exceeds even this highest figure with an 81% incidence of associated anomalies, with over half (56%) with significant cardiac anomalies, which was the leading factor in mortality.

Any reason for these significant differences among twins with tracheoesophageal fistula is not immediately apparent. Certain conclusions can be made with regard to such patients, however. We have not routinely used operative bronchoscopy before thoracotomy for we believe that in the vast majority of patients, no useful information is gained. However, with the foregoing information we would certainly use routine bronchoscopy in all twins with this diagnosis. Also, before definitive operative repair, a cardiac ultrasound is used with a twofold purpose: to ascertain the side of the aortic arch, and to investigate for associated cardiac anomalies.

TABLE 1. *Anatomic Classification*

	No. of Patients (%)
Proximal atresia, distal fistula	10 (63)
Proximal and distal fistulae	1 (6)
Fistula without atresia	3 (19)
Laryngotracheoesophageal cleft	2 (13)

TABLE 2. *Associated Anomalies*

	No. of Patients (%)
Overall	13 (81)
Low birth weight	10 (63)
Cardiac anomalies	9 (56)
Genitourinary/musculoskeletal	6 (38)

The operative repair of esophageal atresia and tracheoesophageal fistula is one of the most challenging problems faced by the pediatric surgeon. Complex anatomy, a right-sided aortic arch, and a higher incidence of associated significant congenital defects may increase even further the risks of dealing with this problem unless adequate preoperative and operative attention to detail is accomplished in the twin.

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